

WHAT IS CLAIMED:

1. A method for treating a disease caused all or in part by a deficiency in *N*-acetylgalactosamine-4-sulfatase comprising the step of administering a recombinant *N*-acetylgalactosamine-4-sulfatase
- 5 2. The method of claim 1 wherein the disease is a mucopolysaccharidosis.
3. The method of claim 1 wherein the disease is MPS VI.
4. The method of claim 1 wherein the disease is Maroteaux-Lamy Syndrome.
- 10 5. The method of claim 1 wherein a patient suffering from the disease demonstrates about 50% or less of a normal *N*-acetylgalactosamine-4-sulfatase activity.
6. The method of claim 1 wherein at least about 50 Units/kg or at least about 1 mg/kg of a recombinant *N*-acetylgalactosamine-4-sulfatase is administered weekly to a patient suffering from a deficiency thereof.
- 15 7. The method of claim 1 wherein at least about 100 units or 2.0 mg/kg of a recombinant *N*-acetylgalactosamine-4-sulfatase is administered weekly to a patient suffering from a deficiency thereof.
8. A pharmaceutical composition comprising recombinant *N*-acetylgalactosamine-4-sulfatase and a pharmaceutically acceptable carrier.
- 20 9. The pharmaceutical composition of claim 8 further comprising a sodium chloride solution, a buffer and human albumin.
10. The pharmaceutical composition of claim 8 wherein the recombinant *N*-acetylgalactosamine-4-sulfatase is present at a concentration of about 1-5 mg/mL or about 50 to about 250 Units per mL.
- 25 11. The pharmaceutical composition of claim 8 wherein the human albumin is present at a concentration of at least about 1 mg/mL.
12. The pharmaceutical composition of claim 8 wherein the buffer is a sodium phosphate buffer at a concentration of about 10-50 mM.

13. The pharmaceutical composition of claim 8 wherein the pH of the solution is maintained at about 5.8.

14. The pharmaceutical composition of claim 8 further comprising polyoxyethylenesorbitan 20 or 80.

5 15. The pharmaceutical composition of claim 14, wherein said polyoxyethylenesorbitan concentration is about 0.001% (W/V).

16. A method for producing a recombinant *N*-acetylgalactosamine-4-sulfatase enzyme comprising the steps of:

- 10 (a) growing cells transfected with a DNA encoding all or a biologically active fragment or mutant of a human *N*-acetylgalactosamine-4-sulfatase enzyme,
- (b) introducing the transfected cells into a bioreactor,
- (c) supplying a growth medium to the bioreactor,
- (d) harvesting said medium containing said enzyme; and
- 15 (e) substantially removing the transfected cells from the said harvest medium.

17. The method of claim 16 wherein the transfected cells are grown on a growth medium comprising a JRH Excell 302 medium supplemented with one or more agents selected from the group consisting of L-glutamine, glucose, hypoxanthine/thymidine and G418.

25 18. The method of claim 16 wherein the transfected cells are grown in a bioreactor for about 5 to 15 days.

19. The method of claim 16 wherein the transfected cells are grown in a bioreactor for about 9 days.

20. The method of claim 16 wherein the transfected cells are substantially 30 separated from the media containing the enzyme through successive membranes.

21. The method of claim 20 wherein the successive membranes are 10 μm , 1 μm or 0.2 μm .

22. A cell line transfected with a DNA operable to produce a recombinant *N*-acetylgalactosamine-4-sulfatase enzyme or a biologically active fragment, analog or mutant thereof; wherein said enzyme is secreted by the cell line or remains in the cell line.

5 23. A cell line according to claim 22 wherein the transfected cell is a Chinese Hamster Ovary cell.

24. A cell line according to claim 23 wherein the transfected cell is a CHO-K1 cell.

10 25. A cell line according to claim 24 wherein the transfected cell is a CSL4S-342 cell.

26. A vector operable to produce a recombinant *N*-acetylgalactosamine-4-sulfatase or a biologically active fragment, analog or mutant thereof.

15 27. A recombinant *N*-acetylgalactosamine-4-sulfatase or biologically active fragment, analog or mutant thereof produced in accordance with the method of claim 16.

20 28. The recombinant *N*-acetylgalactosamine-4-sulfatase or biologically active fragment, analog or mutant thereof having a molecular weight of about 55 to 56 kDa.

29. The recombinant *N*-acetylgalactosamine-4-sulfatase or biologically active fragment, analog or mutant thereof having a molecular weight of about 64 kDa after glycosylation.

25 30. A method to purify a recombinant *N*-acetylgalactosamine-4-sulfatase enzyme or biologically active fragment, analog or mutant thereof comprising the steps of:

- (a) harvesting fluid obtained from a culture of cells transformed with a gene encoding a recombinant *N*-acetylgalactosamine-4-sulfatase or biologically active fragment, analog or mutant thereof;
- (b) running the fluid on a DEAE sepharose column;
- (c) running the fluid on a blue sepharose FF column;
- (d) running the fluid on a copper chelating sepharose column;
- (e) running the fluid on a phenyl sepharose column; and
- (f) diafiltering the purified enzyme.

31. The method of claim 30 wherein the pH of the harvest fluid is adjusted to about 5.0 to 7.3.

32. A method for purifying recombinant *N*-acetylgalactosamine-4-sulfatase comprising the steps of:

- (a) harvesting fluid obtained from a culture of cells transformed with a gene encoding A recombinant *N*-acetylgalactosamine-4-sulfatase or biologically active fragment, analog or mutant thereof;
- (b) running the fluid on a DEAE sepharose column;
- 10 (c) running the fluid on a blue sepharose FF column;
- (d) running the fluid on a copper chelating sepharose column;
- (e) running the fluid on a phenyl sepharose column; and
- (f) diafiltering the purified recombinant *N*-acetylgalactosamine-4-sulfatase or biologically active fragment, analog or mutant thereof.